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Case Report

Acute Type B Aortic Dissection Present as Innominate Vein Compression Syndrome: A Rare Presentation☆

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SUMMARY

The term “innominate (brachiocephalic) vein compression syndrome” was first coined by Wurtz et al in 1989 to describe a patient who presented with “unilateral superior vena cava syndrome.” Here we report a 70-year-old male with comorbidities including hypertension, end-stage renal disease with a left arm arteriovenous fistula for hemodialysis, who presented with left brachiocephalic (innominate) vein compression by a Stanford Type B aortic dissection presenting with left arm edema. The diagnosis was confirmed by venography and computed tomography scan, and he underwent successful endovascular stent grafting.

1. Introduction

The term “innominate (brachiocephalic) vein compression syndrome” was first coined by Wurtz et al[1] in 1989 to describe a patient who presented with “unilateral superior vena cava syndrome.” Here we report a case of left brachiocephalic (innominate) vein compression by a Stanford Type B aortic dissection presenting with left arm edema in a 70-year-old male patient. The patient's comorbidities included hypertension, and end-stage renal disease with a left-arm arteriovenous fistula for hemodialysis.

2. Case report

A 70-year-old male patient with hypertension history presented to the emergency department with a chief complaint of progressing left arm swelling for 10 days, which used to be a site for hemodialysis [brachial-radial arteriovenous (AV) fistula] over the past 6 months. The left arm swelling made hemodialysis difficult and was referred for further survey. On emergency department examination, his vital signs were as follows: blood pressure 224/123 mmHg in the right arm, and a regular heart rate of 73 beats/min. The AV fistula still had thrill on palpation and bruit on auscultation. Chest plain film revealed widened mediastinum (Fig. 1A) and computed tomography demonstrated a dissecting aneurysm approximately 5.2 cm in maximal diameter involving the distal arch and proximal descending thoracic aorta (Fig. 1B) and the existence of an intimal flap at the level above iliac bifurcation. Percutaneous transluminal angiography was performed and we noted the stasis of contrast medium in the brachiocephalic vein with collateral branches enhanced (Fig. 1C). However, there was no critical stenosis or intraluminal thrombus in the brachiocephalic vein during the procedure of balloon angioplasty highly suggesting external compression.

Successful thoracic endovascular aortic repair (TEVAR) was performed with the chimney technique over the left subclavian artery for type B dissecting aneurysm (5.2 cm involving the distal arch and proximal descending thoracic aorta) on the 15th day after admission with a thoracic stent graft (GORE TAG 34 mm × 15 mm; 28 mm × 10 mm; 28 mm × 15 mm) and left subclavian artery stent graft (GORE VIABAHN 13 mm × 50 mm). Left AV fistula regained normal function only after the surgery and hemodialysis was smoothly through the AV fistula on the 16th day after admission, with fairly well controlled blood pressure. Follow up venography 1 week after discharge showed no more contrast medium stasis in the brachiocephalic vein and also no collateral branches enhanced (Fig. 1D).

☆ Conflicts of interest: The authors declare no conflicts of interest in this manuscript.

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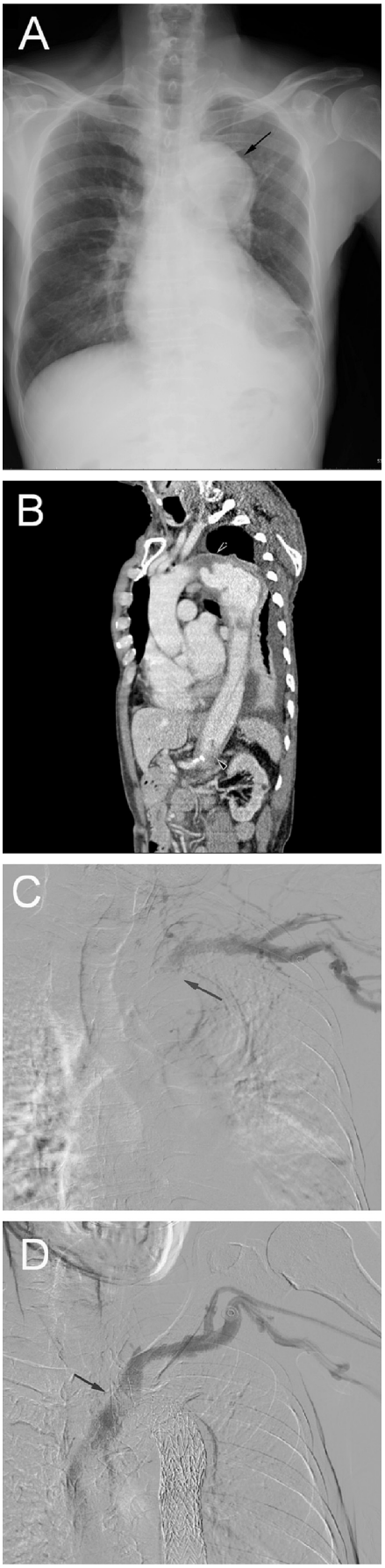

3. Discussion

The classic presentation of aortic dissection includes sudden severe chest, back, or abdominal pain characterized by ripping or tearing discomfort. Only a small fraction of patients present with symptoms secondary to the complications of dissection, as our case will illustrate[2]. The patient's main complaint was progressive left arm edema, which led to difficulties for venous or fistula access to perform dialysis.

Arm edema as the primary finding in aortic dissection, albeit possibly confounded by end-stage renal disease with an AV fistula in the affected arm in this patient, is unusual. It was only on percutaneous transluminal angiography that the left brachiocephalic vein was found to be compressed. Such findings were first described by Wurtz et al[1] as “innominate (brachiocephalic) vein compression syndrome” in a patient whose brachiocephalic vein had been compressed intermittently by the supra-aortic trunks during expiration. Unlike in the original description, the cause of compression of the brachiocephalic vein in our patient was acute aortic dissection with resultant aneurysmal aortic remodeling.

The types of aortic dissection, according to the DeBakey[3] and Stanford[4] classification schemes, are based on the anatomic involvement of the dissection. In the DeBakey classification, aortic dissection is divided into three types: Type I, when the intimal tear occurs in the ascending aorta, whether or not the descending aorta is also involved; Type II, when the tear involves only the ascending aorta; and Type III, when only the descending aorta is involved. In the Stanford classification, aortic dissection is divided into Type A and Type B, which involves either the ascending aorta or the descending aorta, respectively. The Stanford classification helps to guide management, whether primarily surgical or medical.

Based on the anatomical location of the intimal flap, which was located in the descending aorta just proximal to the common iliac bifurcation, our patient suffered a Stanford Type B aortic dissection. However, the dissection proceeded in a retrograde fashion with aneurysmal dilatation of the distal aortic arch and descending thoracic aorta. Retrograde extension of aortic dissection is more commonly associated with Stanford Type A dissections, which are managed differently from Type B dissections. Although the mechanism of propagation of antegrade dissection could be intuitively understood, there are no etiological factors that can be confirmed to predispose to a spontaneous retrograde dissection. However, some iatrogenic retrograde aortic dissections have been described, such as a rare immediate and delayed complication from endovascular aortic stent graft repair[5].

Acute Type B dissections can involve the aortic arch. Compared to simple uncomplicated acute Type B aortic dissections, retrograde extension of the dissection into the aortic arch or ascending aorta is associated with worse survival outcomes compared to dissections restricted to the descending aorta. However, a recent study on Type B dissections found that when such dissections also involve, through retrograde extension, the aortic arch, they are not associated with higher follow-up mortality compared to those without aortic arch involvement[6].

Figure 1. (A) Widening of left superior mediastinum; (B) dissecting aneurysm approximately 5.2 cm in maximal diameter involving distal arch and proximal descending thoracic aorta. The intimal flap end up at the level above iliac bifurcation; (C) contrast medium injected from venous side of the brachial-radial arteriovenous (AV) fistula through an intravenous catheter. Contrast medium stasis in brachiocephalic vein and subclavian vein with enhanced collateral branches; (D) venography after thoracic endovascular aortic repair (TEVAR) with left subclavian chimney stent graft. No contrast medium stasis in the central venous system and also no collateral branches enhanced.

The preferred treatment of many Stanford type B dissections is medical therapy. In our patient with an aneurysmal dilatation of the distal arch, simple medical management alone would be inappropriate[7]. Following adequate medical and preoperative management, a thoracic stent/graft was inserted. TEVAR attracted attention due to its minimal invasiveness for years, and is an indispensable technique for aneurysms at present[8]. Although there are still problems such as a short landing zone, TEVAR with the chimney technique is applied to fix the problem, such as in our case. However, some meta-analyses have showed that it is a viable option for treatment of patients with thoracic aortic pathologies, for emergent as well as elective situations. A higher mortality rate of traditional open surgery compared to TEVAR (10.6% vs. 33.9%) makes it likely to be replaced[9]. The patient's postoperative recovery was uneventful and he was subsequently discharged.

To our knowledge, this is the first reported case of Type B aortic dissection presenting as left-sided upper extremity edema with brachial-cephalic AV fistula dysfunction. This rare case is a reminder that prompt diagnosis and treatment of aortic dissection is critical in preventing consequential morbidity and death.

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